

Definition

The movement disorders include tremor, chorea, athetosis, myoclonus, and asterixis.

Tremor consists of purposeless involuntary movements resulting from the alternating contractions of opposing muscle groups. Tremor at rest occurs when muscles are at rest, for example, the pill-rolling tremor of parkinsonism. Postural tremor occurs when muscles maintain a posture such as outstretched arms, for example, the fine tremor of hyperthyroidism. Action tremor (intention tremor) occurs near the end of a goal-directed movement, for example, the coarse, side-to-side tremor of cerebellar disease seen as the finger-nose test is done.

Chorea is a series of brief, jerky, explosive movements, or "fidgeting"; an example is Sydenham's chorea, seen in rheumatic fever. *Athetosis* is writhing, sinuous movements, especially marked in the digits and extremities, such as occur in hepatic encephalopathy. *Myoclonus* consists of sudden brief twitches or jerks of groups of muscles or a single muscle, as seen in metabolic encephalopathies such as uremic encephalopathy. *Asterixis* ("liver flap") is an intermittency of sustained posture, illustrated by "flapping" of the hands when the arms are outstretched and wrists dorsiflexed, as in hepatic encephalopathy. A "foot flap" is seen in many patients with asterixis of the hands.

Technique

Abnormal movements often can be observed when taking the history or performing parts of the physical examination. If such movements are observed or suspected, they can be studied more carefully by inspecting the patient at rest, while maintaining a posture such as erect with outstretched arms, and performing goal-directed movements such as the finger-nose-finger test.

Systematically inspect the resting patient beginning with the face and proceeding with the upper extremities, trunk, and lower extremities. Chorea produces sudden grimaces about the face—the patient is unable to keep the tongue protruded, and it darts in and out. Inspect the tongue as it rests inside the mouth, as certain types of tremor can involve the tongue. Myoclonic twitches can be seen quite well about the shoulders and distal upper extremities. Restless fidgeting or purposeless movements of the arms are among the earliest signs of chorea. Sinuous twistings of the hand are seen with athetosis.

Have the patient maintain a posture by standing with arms outstretched, slightly flexed at the elbow, and with hands extended at the wrist, as though halting traffic. Look for the following signs. In asterixis the hands "flap," that is, flex briefly at the wrist, then immediately snap back into the extended position. A foot flap can often be brought out by having the patient dorsiflex the feet and maintain them

in that position. The flap is identical to that seen in the hands. This posture is also well suited to bringing out the abnormalities produced by chorea, athetosis, and myoclonus, since they all will produce changes in the position of the limb. Certain tremors are provoked by sustained posture. Observe carefully for distal postural tremors, such as the fine rapid tremor of the hands seen in hyperthyroidism. Tremors involving the shoulders and neck produce head bobbing when the neck is affected and coarse tremors of the entire arm when the shoulder girdle is involved.

Observe voluntary goal-directed movements. Ask the patient to perform the finger-nose-finger test (see Chapter 69, *The Cerebellum*), and watch for tremors that usually occur maximally just before the goal is reached. Note whether the involved segments are distal (wrist, fingers) or proximal (shoulder), as this helps in neuroanatomical localization. A coarse, side-to-side tremor is characteristic of cerebellar disease. Chorea is manifested as a break or interruption in the performance of what is intended to be a smooth voluntary movement.

The following information should be recorded for all abnormal movements:

1. Structures or segments involved
2. State of muscles when they occur: at rest, maintaining posture, or during goal-directed movements
3. Description of movement(s), including the pattern of involvement of various segments and their duration and frequency
4. Factors which increase or decrease movements: rest, exercise, anxiety, alcohol

The progress and influence of treatment on tremors that involve the hands can be followed by having the patient write the same sentence and draw an Archimedes spiral each visit.

Basic Science

The mechanisms underlying involuntary movements are very poorly understood. The extrapyramidal motor system is clearly involved in certain tremors, chorea, and athetosis. This system refers anatomically to the basal ganglia (caudate, putamen, globus pallidus, and amygdala) and related brainstem reticular formation. Experimental evidence also suggests that the ventral lateral nucleus of the thalamus and the cerebral cortex are involved.

Disease of the cerebellum or its brainstem connections produces a coarse action tremor. This tremor has been experimentally reproduced by lesions of the dentate nucleus or the brachium conjunctivum, which contains a large number of projection fibers from the dentate nucleus. Clinically, lesions of the cerebellum or brachium conjunctivum prior to its decussation produce ipsilateral tremor.

Myoclonus is seen in a wide variety of disturbances. An important form occurs in multiple muscle groups in association with metabolic encephalopathy, especially uremic or carbon dioxide encephalopathy. In these circumstances myoclonus presumably indicates neuronal injury. However, the specific pathophysiology is unknown.

Asterixis was described by Adams and Foley in 1949; it is seen in many of the metabolic encephalopathies. Physiologically the electromyogram shows a lapse of electrical activity in the muscle as the wrist flaps down, followed by a compensatory muscle contraction that jerks the hand up again. The neurophysiology is not known.

Clinical Significance

Tremor

The pill-rolling tremor of parkinsonism is present at rest. This is a coarse, regular movement involving the thumb and index finger. Although the tremor predominantly involves the distal upper extremity, the face and tongue may be involved also. Sleep decreases the tremor, and emotion makes it worse. The tremor often increases as the patient walks.

Other causes of tremor at rest are rarer. They include drug-induced tremors (principally phenothiazines), severe cases of essential tremor (benign familial tremor), Wilson's disease, chronic acquired hepatocerebral degeneration, mercury poisoning, and general paresis.

As a generalization, proximal postural tremors—those involving the shoulder, pelvic girdle, or neck—are produced by lesions of the cerebellum and its brainstem connections. They are usually coarse and slow. Distal postural tremors, involving wrists and finger joints, result from mid-brain or basal ganglia problems. Many of these are fine and rapid. However, there are exceptions to these generalizations, and in many cases the site of neurologic dysfunction is not really known.

The most common postural tremors are as follows:

1. Anxiety and fatigue tremors: fine, rapid tremors involving the fingers. The injection of epinephrine into normal individuals produces tremors identical to those seen in anxiety or with fatigue.
2. Thyrotoxicosis: a fine, rapid tremor of the fingers identical to the tremors listed above.
3. Essential (benign familial) tremor: a coarse, irregular tremor that usually starts in the hands and fingers and eventually involves the voice, head, and neck. Usually it becomes worse with goal-directed movements. Difficulty with handwriting, piano playing, typing, or drinking coffee is often the most prominent complaint. Drinking alcohol diminishes the tremor. There is a strong heritable tendency. Onset is in early adult life. Treatment with propranolol is efficacious in some patients.
4. Cerebellar tremor: a coarse, irregular tremor involving the shoulder girdle and neck, seen in patients with diseases of the cerebellum. A prime example is alcoholic cerebellar cortical degeneration. When quite severe, it can be present at rest also.
5. Lithium tremor: seen in individuals being treated with lithium. It is quite similar to essential tremor.

Postural tremors are also seen in Wilson's disease, acquired hepatocerebral degeneration, and certain poisons, espe-

cially mercury. Mercury poisoning was known as the "hatter's shakes" because workers involved in the manufacture of felt hats were exposed to mercury.

Action (intention) tremors are seen during voluntary activity, usually being most prominent near the end of goal-directed movement. The same generalizations about localization apply as given for postural tremors. A good example is the coarse, side-to-side tremor seen in cerebellar disease as the finger approaches the nose. Note that although we speak here of the finger, the actual origin of the tremor is in the elbow or shoulder joint.

The foregoing statements about tremors are generalizations to which there are numerous exceptions. However, they form a useful bedside approach to the analysis of tremor disorders. The final diagnosis is based on this analysis in addition to a meticulous history and physical examination.

Chorea and Athetosis

Sydenham's chorea is seen in children and adolescents and is often associated with rheumatic fever. Huntington's chorea is an autosomal dominant disorder that usually appears in adulthood. Chorea gravidarum is seen in pregnancy. Chorea has been reported in a large variety of other conditions, especially diseases such as lupus erythematosus in which there is cerebral arteritis.

Athetosis occurring during childhood involves both sides of the body ("double athetosis") and is thought to be due to neonatal cerebral hypoxia. In adults it usually involves only one side of the body and is seen most often with strokes.

Myoclonus and Asterixis

Myoclonus involving multiple muscle groups most frequently occurs in patients with metabolic encephalopathies, such as those produced by uremia, carbon dioxide, or hypoxia.

Asterixis was first described in hepatic encephalopathy. It is also seen in other metabolic disorders such as uremia, hypokalemia, carbon dioxide narcosis due to chronic pulmonary disease, dialysis dementia, hypoxia, and hypomagnesemia. Asterixis has been caused by the following drugs: dilantin, phenobarbital, carbamazepine, primidone, and perhaps diazepam and flurazepam. Structural lesions producing asterixis, even unilateral asterixis, have been located in the midbrain and thalamus.

The combination of myoclonus and asterixis in a patient with stupor strongly suggests a metabolic encephalopathy, rather than a structural brain lesion, as the cause.

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